APPENDIX A

***Abbreviations Table A-C**: NH, natural history, AV, atrio-ventricular, VA, ventriculo-arteriel, HLHS, hypoplastic left heart syndrome, AA, aortic atresia, ToP, termination of pregnancy, eHLHS, evolving HLHS, BV, biventricular, UV, univentricular, IAS, intact intra atrial communication, Htx, heart transplantation, GA, gestational age, BW, birth weight.*

***Table A**. The studied cohorts in Paper I-V with inclusion and exclusion criteria and description of the cohort.*

Paper		Cohorts and subgroups	n	Inclusion criteria	Exclusion criteria
I					
	Cohort I	Infants with single ventricle physiology and a shunt as the only source of pulmonary blood flow	28	Surgery from September 2007-December 2010	Lack of consent
	Cohort II		49	Surgery from August 2002-June 2007	-
II					
	Cohort NH	Fetuses with aortic stenosis 2005-2012 in ongoing pregnancies	107	Situs solitus, AV/VA concordance, stenosed but patent aortic valve	Fetal intervention
	Subgroup 1	Postnatal treatment and intention-to-treat	85	Available fetal echo data ≤ 30 completed weeks	
	Subgroup 2	Boston 2006 criteria for UV circulation	44		
	Subgroup 3	Boston 2009 threshold score for intervention	12		
III					
	Cohort NH	Paper II	107	"	Fetal intervention
	Cohort FV	Fetuses with aortic stenosis 2005-2012 in ongoing pregnancies	67	Fetal intervention, intention to treat	-
	Subgroup 1	Successful FV	59		
	Subgroup 2	Unsuccessful FV	8		
	Subgroup 3	Hydrops	24		
	Propensity score and IPTW matched cases	NH and FV cohorts matched on propensity score	65	Available fetal echo data ≤ 30 completed weeks Liveborn neonate with known outcome	-
IV					
	Cohort HLHS/AA	Neonates with HLHS/aortic atresia born in Sweden 1990-2010	254	Liveborn neonates	Aortic stenosis. Other cardiac malformation.
V					
	Cohort HLHS/AA, surgery	Neonates with HLHS/aortic atresia born in Sweden 1990-2010	121	Surgery 1993-2010	Aortic stenosis. Other cardiac malformation.

Table B. *The outcomes, exposures and possible confounding factors and interactions observed in the cohorts with fetal aortic stenosis, Paper II and III.*

Fetuses with prenatal diagnosis of aortic stenosis		
Outcome (y)	**Exposures (x)**	**Confounding and interactions**
ToP	Prenatal diagnosis of fetal aortic stenosis	Prenatal cardiac screening in the population Legislation Cultural and religious aspects Known criteria for eHLHS versus likely BV repair Possibility to treat in utero Possibility to treat after birth
sIUD	Cardiac failure	Other causes of cardiac failure
Fetal intervention	Cardiac morphology, size and physiology	The option of ToP Availability and attitudes towards fetal intervention
Liveborn neonates with prenatal diagnosis of fetal aortic stenosis		
Outcome (y)	**Exposures (x)**	**Confounding and interactions**
Comfort care	Parental wish	Additional malformations Legislation
Neonatal death before first procedure	Circulatory failure	Other causes of circulatory failure Planning of delivery
Liveborn neonate with prenatal diagnosis of fetal aortic stenosis and postnatal procedure performed		
Outcome (y)	**Exposures (x)**	**Confounding and interactions**
BV circulation **BV to UV circulation** **UV circulation**	Prenatal diagnosis of fetal aortic stenosis	Gestational age at delivery Birth weight Cardiac morphology, size and physiology at birth Birth location Surgical center
30-day mortality	All postnatal procedures	IAS, sepsis, prematurity, cardiac or respiratory failure
Overall survival	BV or UV circulation	Type and number of surgical procedures Cardiac or respiratory failure Pulmonary hypertension Htx (none)

Table C. *The outcomes, exposures, possible confounding factors and potentially modifiable factors in the observed cohorts with HLHS/AA, Paper IV and V.*

HLHS/AA, liveborn neonates			
Outcomes (y)	**Exposures (x)**	**Confounding factors**	**Modifiable factors (x)**
Surgery versus no surgery	Gender Birth period Prenatal diagnosis GA ≤ 37 weeks BW ≤ 2500 g Birth location Extra cardiac malformations	Experience of the surgical treatment	Legislation Ethical considerations Information to parents Attitude towards comfort care among professionals and in the society
HLHS/AA, surgery			
Outcomes (y)	**Exposures (x)**	**Confounding factors**	**Modifiable factors (x)**
30-day mortality **Inter stage mortality**	Birth period	-	Prenatal diagnosis Experience Surgical modifications Anti-coagulation therapy Home monitoring
Overall survival	Gender Birth period Prenatal diagnosis Gestational age at birth Somatic growth at birth Extracardiac malformations Surgical center Age at first surgery	Prenatal diagnosis Experience Surgical modifications Anti-coagulation therapy Home monitoring	Experience Prenatal diagnosis Gestational age at birth (prenatal diagnosis) Surgical center Age at first surgery Follow-up